

Kronik tromboembolik pulmoner hipertansiyon nedeniyle uygulanan iki taraflı pulmoner tromboendarterektomi: Bölgemizdeki en küçük olgu

Bilateral pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension: the youngest case in our region

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Özet- Hidrosefali tedavisi için ventriküloatriyal (VA) şantların kullanılması kronik tromboembolik pulmoner hipertansiyon (KTEPH) gelişimi ile birliktedir. Kronik tromboembolik pulmoner hipertansiyon kronik veya tekrarlayan emboliler sonrası gelişir. Çocuklarda nadir görülmekle birlikte tahrip edici potansiyeli yüksektir. Pulmoner tromboendarterektomi (PTE) KTEPH'li hastalarda önemli ve iyileştici bir tedavi yöntemidir. Bu yazıda, VA şant işlemi sonrası KTEPH gelişen 14 yaşında bir erkek olgu sunuldu. Hastada başarılı PTE sonrasında sistolik pulmoner arter basıncı 75 mmHg'den 30 mmHg'ye geriledi. Çocukluk çağı KTEPH'de PTE'yi öneriyoruz.

Summary- The insertion of ventriculoatrial (VA) shunts for the treatment of hydrocephalus is associated with the development of chronic thromboembolic pulmonary hypertension (CTEPH). Chronic thromboembolic pulmonary hypertension occurs in patients with recurrent or chronic pulmonary embolism, and is a rare but, potentially devastating disease in children. Pulmonary thromboendarterectomy (PTE) is an important curative therapy for patients with CTEPH. Herein, we present a case of a 14 year-old male patient with CTEPH that developed after a VA shunt procedure. After successful PTE, systolic pulmonary artery pressure was decreased from 75 mmHg to 30 mmHg. PTE is recommended in the pediatric CTEPH population.

Abbreviations:

INR International Normalization Ratio

CTEPH Chronic thromboembolic pulmonary hypertension

PTE Pulmoner tromboendarterectomy

VA Ventriculoatrial

VP Ventriculoperitoneal

Chronic thromboembolic pulmonary hypertension (CTEPH) is a relatively rare, but generally curable cause of pediatric cases of pulmonary hypertension.[1] Sustained activation of coagulation factors, and brain thromboplastin found in the cerebrospinal fluid as a consequence of catheter-related microthrombi, and shunt infection plays an important role in its pathogenesis.[2] In untreated cases, right heart failure, and death can be seen.[3,4] Together with hemodynamic improvement, significant improvements have been achieved in blood gas values, functional capacity results, and survival rates after pulmonary thromboendarterectomy (PTE) .[5] Predisposition to coagulation, history of thromboembolism, and presence of an intravenous catheter in symptomatic patients highly suggest diagnosis of CTEPH[6]

Herein, we present a patient who consulted to our clinic with complaints of dyspnea, and exhaustion following ventriculoatrial (VA) shunt operation, and diagnosed as CTEPH. In this case PTE yielded satisfactory results.

CASE PRESENTATION

A -11-year old male patient consulted to our clinic with complaints of dyspnea, and prostration. The patient had undergone ventriculoperitoneal (VP) shunt procedure for his hydrocephalus when he was 4 years old.. His dysfunctional VP shunt had been replaced by a VA shunt.

However when he was 6 years of age, VA shunt had become dysfunctional, and removed.

On cardiac examination, regular heart sounds with stiff S2 sound without any murmur were heard. Telecardiograms demonstrated a prominent pulmonary conus, and a cardiothoracic index greater than 0.5. On EKG, sinus rhythm at a heart rate of 75/min, right ventricular hypertrophy, and complete right bundle branch block were noted. Echocardiographic examination revealed increased flow rate within the superior vena cava and right atrium junction, signs of pulmonary hypertension, and dilated pulmonary arteries especially on the right side (Figure 1a). During cardiac catheterization pulmonary artery pressure (83/50-67 mmHg), pulmonary resistance (6.3 Wood Unit/m²) , and systemic resistance (37.7 Wood Ünite/m²) were measured. Computed tomography revealed a main pulmonary artery with increased diameter (45 mm). Any thrombus within the cardiac chambers was not detected on cardiac magnetic resonance images. Results of the tests revealing predisposition to thrombosis such as tests for lupus anticoagulant, anticardiolipin antibody, protein C, and S, antithrombin III, factor V Leiden, and prothrombin gen defects were found to be within normal limits. Bosentan treatment was started on the patient, however because of progression of CTEPH, indication for PTE

was established, and performed as described previously (Figure 1b). During the procedure, Jamieson type I, and II lesions were excised, and removed from within right, and left pulmonary arteries, respectively. For PTE, a total of 29 minutes of deep hypothermia was required to achieve circulatory arrest. Intravenous heparin was initiated within postoperative 6 hours.

Postprocedural thrombotic prophylaxis was maintained with warfarin therapy while sustaining therapeutic international normalized ratio (INR). Assisted ventilatory support was withdrawn on postoperative first day. However on the fourth day cardiac tamponade

developed, and re-sternotomy was performed to evacuate accumulated blood collection. On the postoperative ninth day, requirement for the intensive care support was relieved, and on the 15th day his systolic pulmonary artery pressure regressed down to 30 mm Hg, and consequently the patient was discharged. On the control visit at the third postoperative month, any sign of pulmonary hypertension was not detected. Presently, the patient is monitored under prophylactic anticoagulant therapy.

DISCUSSION

Pediatric age group of patients constitutes a small percentage (1.1 %) of CTEPH patients for whom pulmonary endarterectomy is recommended.[1,8] CTEPH is probably overlooked in general population. VA, and VP shunts are used to control hydrocephalus. As a serious complication, pulmonary hypertension is a result of recurrent thromboembolisms.[9] Madani et al [8] recently published the largest series of pediatric cases who had undergone pulmonary endarterectomy with the indication of CTEPH. In their study consisting of 17 cases, 67 % of the patients had a thrombophilic disease, and in only one patient “VA shunt” was determined as a risk factor. Evaluation of hypercoagulability in CTEPH conveys a great importance as for the monitorization of the patients. In this study, as is the case in our patient, any risk factor besides VA shunt could not be determined, and any evidence of related to hypercoagulability could not be found. In the same study, the most frequent postoperative complication of PTE was recurrent thrombosis detected in 38 % of the patients which underlines the importance of anticoagulation. Our patient is still monitored closely as for development of thrombosis, and receiving

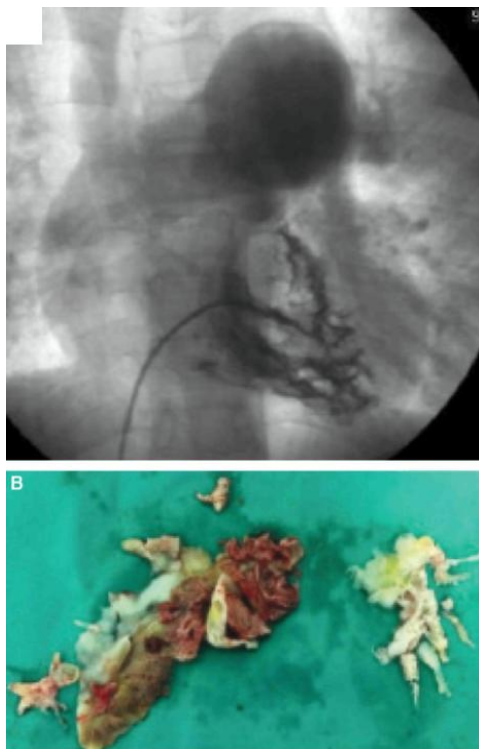


Figure . (A) Angiographic images of the right ventricle, and pulmonary arteries.. **(B)** Macroscopic appearances of type 1 (*right*), and II (*left*) mass lesions excised, and extracted from within the pulmonary artery during endarterectomy procedure..

warfarin therapy to sustain an INR level of 2-3.

Our case is the youngest patient who had undergone pulmonary endarterectomy in our country who completely recovered without any postoperative late-term complication. Our patient is monitored meticulously for the recurrence of thrombosis.

Ventriculoatrial shunts carry important risks as for thromboembolism, and pulmonary hypertension. Therefore these cases should be carefully monitored. PTE achieves satisfactory results in cases with pulmonary hypertension secondary to chronic pulmonary thromboembolism.

Conflict of interest: None declared

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Anahtar sözcükler: Antihipertansif ilaç/terapötik kullanım; çocuk; endarterektomi/yöntem; hipertansiyon, pulmoner/etioloji; kronik hastalık; tromboemboli/komplikasyonlar.

Key words: Antihypertensive agent/therapeutic use; child; endarterectomy/method; hypertension, pulmonary/etiology; chronic disease; thromboembolism/complications.